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Case Report

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Osteoskeletal Manifestation of Scurvy in a Male Infant (Case Report)

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Abstract

Scurvy, a disease of dietary deficiency of vitamin C, was uncommon in the last century due to improved nutrition and health care. However, in the last decade it was increasingly reported in adult population. In a period of infancy, scurvy appears after the age of five months and is related mainly to malnutrition. The main symptoms are vascular purpura, bleeding and gum abnormalities, but musculoskeletal manifestations could also be prominent. Children experience severe lower limb pain related to subperiosteal bleeding. The misdiagnosis with rickets and arthritis is common. We present a case of scurvy in 8 month boy with osteoskeletal manifestation, very rare seen in contemporary European pediatrics. The diagnosis of our patient is made by clinical presentation (such as haematuria, edema of lower extremities as well as swollen right leg), some biochemical parameters and radiological finding. The therapeutically outcome and follow-up of the boy confirmed the diagnosis and etiology.

Introduction

The vitamins are naturally occurring organic substances which possess unique molecular structures that the organism is unable to synthesize, and consequently must be supplied trough the food. The morphological changes of the vitamin deficiency reflect the injury caused by defective chemistry of cells. The kind of injury may be peculiar to one type of cell or more generally distributed in several tissues depending to biochemical process affected by the deficiency. Vitamin C is water-soluble substance, very sensible to heat, ultraviolet or oxygen exposure. The total pool in the body is 1500-2500 mg. Absorption occurs in the ileum. Vitamin C is not stored in the body, so the dietary intake is indispensable. To prevent scurvy the daily intake must be minimum 10 mg/day (1, 2). The classic clinical and pathologic features of infantile scurvy were firstly described by Thomas Barlow in 1883 (3, 4). In modern western society, scurvy very rarely occurs in adults, although infants and elderly people could be affected. Vitamin C is destroyed by the process of pasteurization, so babies fed with ordinary bottled milk sometimes suffer from scurvy if they are not provided with adequate vitamin supplements (5, 6). Scurvy is caused by a prolonged deficiency of vitamin C intake that results in defective collagen synthesis, tissue repair, and synthesis of lipids and proteins. It functions, both as a reducing agent and as an antioxidant, are necessary for many physiologic functions, including metabolism of iron and folic acid, resistance to infection, and integrity of blood vessels.

Vitamin C is required for prolyl and lysyl hydroxylase activity and is essential for collagen synthesis. Defective collagen composition compromises skin, joint, bone, and vascular integrity. Vitamin C is also required for carnitine synthesis, and it is critical for fatty acid transportation into the mitochondria. This oxidative metabolism is also critical for muscle function (6,7).

Scurvy occurs after vitamin C has been eliminated from the diet for at least 3 months and when the body pool falls below 350 mg (8,9).

Symptoms and signs of scurvy may be remembered by the 4 Hs: hemorrhage, hyperkeratosis, hypochondriasis, and hematological abnormalities. Bleeding into the joints causes exquisitely painful hemarthroses. Subperiosteal hemorrhage may be palpable, especially along the distal portions of the femurs and the proximal parts of the tibias of infants. In advanced cases, clinically detectable beading may be present at the costochondral junctions of the ribs. This finding is known as the scorbutic rosary. Bleeding into the femoral sheaths may cause femoral neuropathies, and bleeding into the muscles of the arms and the legs may cause woody edema. Anemia develops in 75% of patients, resulting from blood loss into tissue, coexistent dietary deficiencies (folate deficiency), altered absorption and metabolism of iron and folate, gastrointestinal blood loss, and intravascular hemolysis. The anemia is most often characterized as normochromic and normocytic (9,10).

Scurvy commonly occurs in infants between ages 5–24 months, with a peak between 8–11 months (5,9,10). In a more recent review of cases of scurvy in Thailand 93% of reported cases were in children between 1–4 years of age, although cases were reported in individuals as young as 10 months and as old as 9 years (11). Between 1997–2001, Ortner et al. (12,13) carried out various investigations of pathological features related to abnormal porosity of the cortex. They suggested that the pathological lesions identified are a response to chronic bleeding at the site of the porosity or hyperplasia, and related such abnormal bleeding to scurvy.

Early clinical manifestations of scurvy consist of pallor, irritability, and poor weight gain. In advanced infantile form, the major clinical manifestation is extreme pain and tenderness of the arms and, particularly, the legs. The baby is miserable and tends to remain in a characteristic immobilized posture from subperiosteal pain, with semi flexion of the hips and the knees.

Some authors have suggested that the most characteristic diagnostic radiological finding of vitamin C deficiency is a large, fluctuant, parietal swelling, which is apparently caused by subperiosteal hemorrhage. However, long bone changes are better clinical identifiers of vitamin C deficiency than parietal swelling. The epiphyses and periosteum also become easily detachable because of hemorrhage below the periosteum. Separation of the metaphyseal plate from the diaphysis, epiphyseal clefts, and misalignments of the metaphysis may also occur. A circular, opaque radiological shadow often surrounds epiphyseal centers of ossification. This ring of increased opacity formed around the ossification center of long bone epiphyses is known as the Wimberger sign, which may result from bleeding or attachment movement (14,15,16). The goal of treatment of scurvy is to saturate the body rapidly with ascorbic acid. At maximum doses, body stores become saturated in a few days.

The aim of this report is to present a case of scurvy in 8 month boy with osteoskeletal manifestation, very rare seen in contemporary European pediatrics.

Case report

The patient we present is K.P., 8 month old boy, the 12the born in a poor family, where one previous child died from malnutrition and dehydration in the period of infancy. K.P. was fed only with cow milk and biscuits. No vaccination and anti-rickets prophylaxis were applied. From the period of five months the boy manifested irritability, sweating, poor appetite and cried when somebody touched him. The main problems which emerge the hospitalization were haematuria, edema of lower extremities as well as swollen right leg.

At admission the baby was a febrile, pale, malnourished, without purpura, his hair was dry and cracked. The anthropometric measures showed high and weight on the 3rd percentile. Clinical evaluation showed no organomegaly, no neurological signs, no gingival bleeding and only one tooth was present. Laboratory findings are presented on the Table 1.

Table 1: Laboratory findings (at admission and during the treatment).

Parameter	Values				Reference Value
Hemoglobin (g/L)	58	88	97	115	90-140
Erythrocytes ($x10^{12}$)	3.5	4.33	4.33	4.93	3.1-4.5
Leucocytes ($x10^{9}$)	19.7	14.7	8.8	9.3	6-15
Platelet s($x10^{9}$)	264	364	248		> 50
Serum iron (nmol/L)	2.2			10.5	7.16-17.90
Urine	Blood ++	Blood +	Negative		
Urea (mmol/L)	3.4				119-327
Creatinin (mmol/L)	19				71-195
Sodium (mmol/L)	132				139-146
Potassium (mmol/L)	4.4				4.1-5.3
Calcium (mmol/L)	2.24				2.20-2.70
Phosphorus	1.12				1.4-2.1
Alkaline phosphatase (IU)	333				50-155
Total serum protein/albumin (g/L)	56/31				62-80/40-50

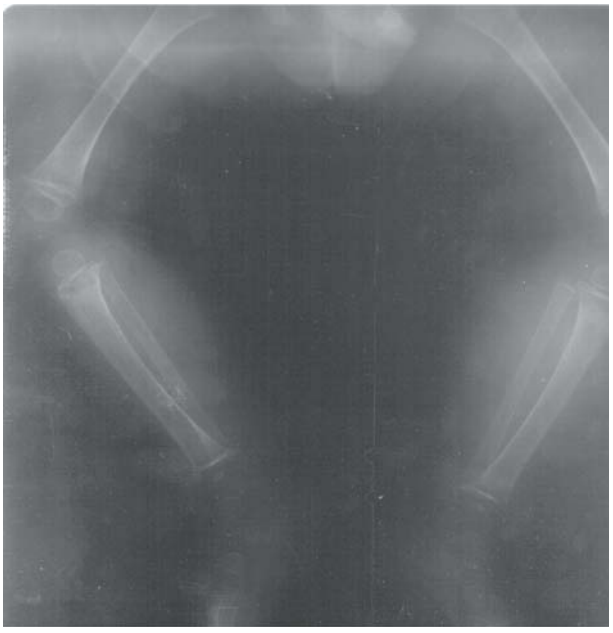

Fig. 1: Edema of the leg, osteoporosis, ricket, no haematoma.

Liver functional tests and hemostasis were in the normal range. Ultrasound of kidney was normal. Doppler of blood vessels of both legs was normal which excluded trombophlebitis. Swelling of the right leg indicated radiological investigation. Massive subperiostal haematoma on the right femur, dilatated metaphises, general osteoporosis, epiphysiolisis and dislocation of the epyphisis have been present on the radiogram (see Fig. 1).

Considering the osteoskeletal manifestation, malnutrition, anemia, irritability and the anamnesis of poor nutrition, as well as the radiological findings, we concluded deficiency of vitamin C. Unfortunately, our laboratory could not measure the level of serum ascorbic acid. The therapeutic interventions comprised erythrocyte transfusion, fresh plasma, large doses of ascorbic acid, vitamin D, B-vitamins and correction of nutritional intake.

Two months later the boy became stable, the edema of the leg disappeared the radiological finding showed organization of the hamatoma (Fig.2a and 2b) and he was dismissed from the hospital. Parents did not come to the arranged control. Bronchopneumonia with fever, and loss of appetite were the indication for the second hospitalization. Radiological control of the leg showed reduction of the subperiostal hematoma (Fig. 3). Antibiotic therapy and another supplementation of ascorbic acid and vitamin D stabilized the general condition of the baby. Until this report, the baby was not presented for control.

In summary, we present a case of scurvy in a male 8 month baby, due to malnutrition, a very rare condition in contamporary European pediatrics. The most typical findings were radiological presentation of musculoskeletal bleeding. Differential diagnosis was:

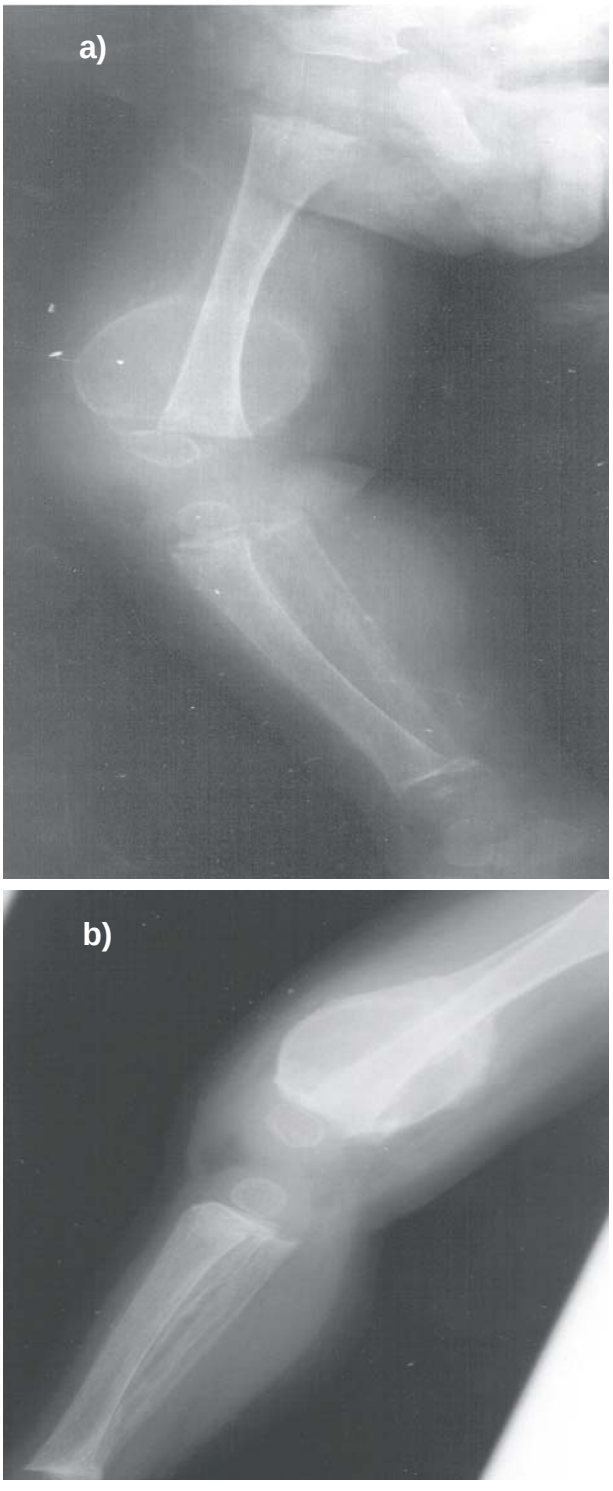

Fig. 2: a) Subperiostal haematoma; b) Dislocation of the epiphysis.

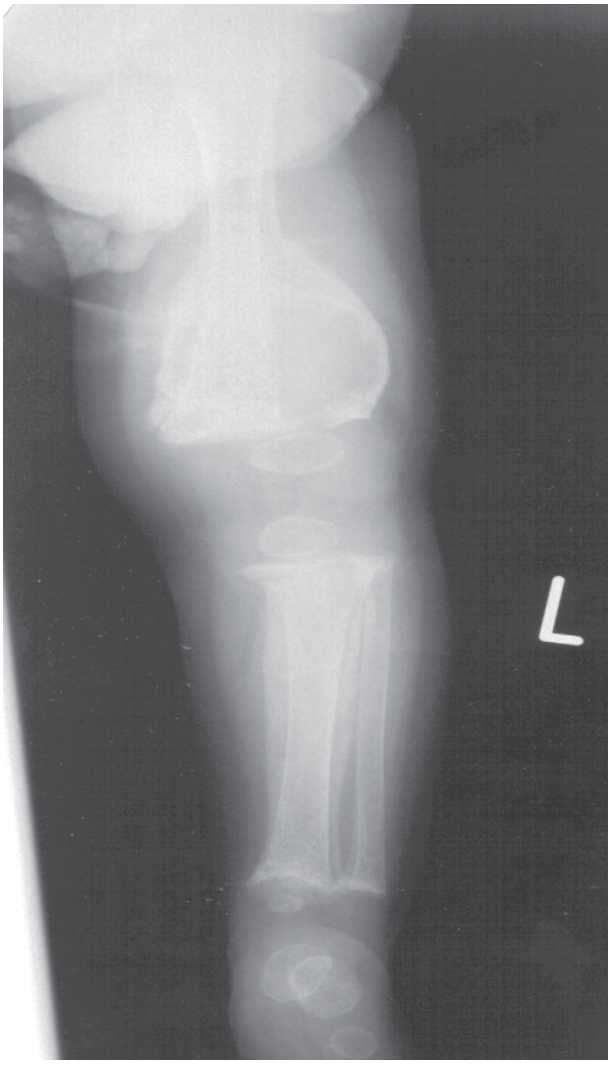

Fig. 4: Organisation of the subperiostal heamatoma.

trauma of legs and joints, deep vein thrombosis, infection or coagulation abnormalities. Supplementation of vitamin C and other vitamins as well as the correction of nutrition are shown to be an effective therapeutic choice.

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